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Decision-making challenges in children with congenital and acquired monorchism: a critical literature review

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Abstract: Monorchism in children can be caused by congenital and acquired conditions, and can potentially influence the hormonal and reproductive function of an individual in the long term. Depending on the etiology, different approaches to the solitary testis have been suggested; however, studies on this topic are scarce. Prevention of anorchia is the main goal in the management of a child with monorchism. The risk of bilateral testicular loss must be weighed against the risk of performing surgery on a healthy gonad. Little is known about the long-term consequences of the various methods for fixation of the testis. This paper provides an up-to-date summary of the current literature on congenital and acquired monorchism in childhood.

Key words: monorchism, children.

Introduction

Monorchism can be congenital or acquired in origin. Regardless of the reason for testis absence, whether it is due to testis agenesis, tumor or torsion, the patient will be confronted with only one functioning gonad. With regard to monorchism, it is important to address two main issues: 1) what is the underlying pathology that led to the testicular loss, and 2) what is the risk that the same process will destroy

the other gonad. Furthermore, special attention must be paid to the contralateral counterpart. Preserving a healthy contralateral testicle is crucial for the patient's fertility and hormonal well-being. However, it remains unclear whether there is a need for contralateral testicle fixation and, if yes, when and how to carry out the procedure. Clinicians currently behave in accordance with their own experience, either performing orchidofixation or leaving the contralateral gonad without any intervention. Due to the lack of strict recommendations in the available guidelines and publications, this article presents a summary of the current knowledge on this topic.

Prevalence

There is no epidemiological data on the incidence of congenital monorchism. Therefore, it can only be estimated based on the incidence of unilateral cryptorchidism, which is seen in 3–6% of all male newborns [1]. At 1 year of age, approximately 1% of boys requires surgical exploration due to non-scrotal testis (undescended testis, UDT). A non-palpable testicle is present in 10–20% of these cases, and vanishing testis syndrome (VTS) is detected in 35–60% [1]. Testicular agenesis is a very rare condition that is sometimes misdiagnosed as VTS.

It is even more difficult to estimate the prevalence of acquired monorchism among prepubertal boys, mainly due to the broad spectrum of pathologies that can cause testicular loss (torsion, trauma and oncological diseases are the most common). The available data is heterogenous and scarce.

Testicular torsion in children is not a rare finding. In the population of young males under 25 years of age, the annual prevalence of testicular torsion varies from 2.9 to 3.8 cases per 100,000 people [2–4]. The highest incidence is in adolescents (12–18 years) and in neonates [5, 6]. The overall testicular salvage rate in young adults (under 25 years) is approximately 70% [3, 4]; however, a necrotic testis is often found during the perinatal period [2].

Testicular neoplasms are typical for adolescents and young adults, with a peak prevalence in males entering their fourth decade of life [7]. Although the incidence varies between countries and races [8], pre-adolescent boys presenting with testicular tumors are very rare, accounting for about 1% of all pediatric solid tumors [9]. Some of these tumors are benign, and a testis-sparing procedure is preferred nowadays.

Epidemiological data regarding testicular trauma in children are scarce. It usually occurs in patients between 6 and 10 years old [10]. The incidence of complications after the injury is believed to be low, and the testis can usually be preserved.

The impact of inflammatory diseases (e.g., mumps) on the testis remains unclear. Even if the gonadal tissue becomes involved during the course of infection, it does not always lead to unilateral testicular loss [11, 12].

Congenital conditions

Nowadays, there is a consensus among most pediatric urologists that the majority of congenital monorchism cases are caused by vascular pathologies, which can lead to unilateral gonadal regression (also called VTS) [1, 13]. It is possible that thrombosis or torsion of the spermatic cord vessels during the fetal and/or perinatal period can result in testicular damage, eventually causing unilateral gonadal atrophy [14, 15]. A solitary testis can also be found in individuals with disorders of sex development (DSD) and metabolic diseases, but it is unlikely that endocrinopathy itself can cause unilateral congenital monorchism [16]. The hormonal excretory function in men with congenital monorchism does not significantly differ to healthy individuals with both testes, provided the remaining testicular tissue has normal morphology [17]. Testicular agenesis, meaning that no testis developed on one side, is believed to be more than two-times less frequent than vascular pathologies [13, 18].

If a child presents with a congenital monorchism, fixation of the contralateral gonad can be considered. This is performed to reduce the risk of torsion of the solitary testis later in life, thereby avoiding a total loss of excretory and reproductive function. On the other hand, the risk of surgery and efficacy of the procedure must be weighed against the possible benefits. The overall risk of complications and the potential need for additional surgical interventions must be explained to the family, and the risk factors for contralateral testicular loss (torsion) should be individually estimated.

In 2014, Martin *et al.* studied the frequency of pathological deformation of tunica vaginalis, the so-called “bell clapper anomaly”, which is believed to be responsible for the majority of cases of postnatal testicular torsion [19]. Their study included 50 patients with perinatal testicular torsion and 27 with acute testicular torsion, and the anatomy of the contralateral testicle was assessed in both groups. In the congenital monorchism group, only one case (2%) presented a partial bell clapper anomaly, whereas a complete bell clapper anomaly was found in 21 of the 27 contralateral testes (78%) in the acute torsion group. The authors suggested that in boys with congenital monorchism, the risk of torsion of the contralateral gonad during their lifetime is too low to justify orchidofixation of the unaffected testis [20]. This conclusion is supported by the fact that the mechanisms of testicular torsion in the perinatal period (spermatic cord together with the tunica vaginalis) is different to that during puberty and in young adults (spermatic cord only within the tunica vaginalis) [21].

The aforementioned approach seems to have become more popular among pediatric urologists and surgeons over time. Nevertheless, for many years, orchidfixation of the contralateral testicle in patients with congenital monorchism was considered the gold standard treatment rather than the exception. This is based on data from a few small series published in the 1980s and 1990s. In 1982, Harris *et al.* showed

that out of 15 patients with congenital monorchism, the bell clapper anomaly was present in the contralateral testicle of 13 (87%) patients. Highlighting that pathologies of the gubernaculum are associated with most instances of antenatal and postnatal torsion, and that those conditions are usually bilateral, the authors proposed surgical exploration and orchifixation as a routine procedure whenever monorchism was diagnosed [22]. In 1985, Bellinger similarly confirmed the occurrence of the bell clapper deformity in 83% of boys with congenital monorchism, also advising contralateral fixation as a routine procedure [23]. Caesar and Kaplan showed a 12% incidence of the bell clapper anomaly on cadavers. The high prevalence of this pathology, which is many times higher than the incidence of testicular torsion, suggested that the bell clapper anomaly is an additional risk factor rather than the sole cause of testicular torsion [24].

In his retrospective study from 2006, Al Zahem *et al.* investigated 31 boys with a confirmed VTS who were subjected to the contralateral orchidodixation for the presence of any anomalies related to the remaining testicle [25]. Testicular abnormalities were found in 22 cases (71%), and among them, five patients (16%) had pathologies that could result in testicular torsion. Overall, there was no evidence of any morbidity associated with the surgical intervention, and no cases of testis torsion were observed during follow up. Hence, the authors recommended routine orchidofixation as a safe and effective procedure in patients with VTS, but also noted the limitations of their work which included only a small number of participants.

In 2018, Monteilh *et al.* published a meta-analysis on the management of neonatal testicular torsion [26]. This study, representing the most recent and up-to-date analysis, not only validated the presumption of a higher frequency of the extravaginal mechanism than the intravaginal one (96.5% vs. 3.5%), but also showed that early bilateral exploration and contralateral testicle fixation can benefit 8–12% of patients. This is due to the high risk of bilateral torsion among neonates. However, this conclusion should not be extrapolated to VTS, as VTS can occur as a result of any vascular accident during fetal life when the testis is on its way to the scrotum.

Acquired conditions

It is believed that the vast majority of postnatal testicular torsion cases have their roots in the presence of a bell clapper anomaly. This is a congenital malformation of the parietal layer of the tunica albuginea that does not adhere to the anterior testicular wall, leaving the testicle and spermatic cord mobile. This condition is often bilateral, and when explored due to torsion of the contralateral testis, the frequency in the healthy testis was found to be as high as 78% [20]. Conversely, in infants operated on due to torsion, the prevalence of this deformity is only about 2%. Such a huge

difference is difficult to explain, and there is no reliable data on the incidence of the bell clapper anomaly in healthy males.

The bell clapper anomaly is thought to be an independent risk factor for intravaginal testicular torsion. Many other conditions, such as temperature [27, 28], the length of mesorchium, history of cryptorchidism [19] or extensive exercises and bicycle riding [6], might also increase the risk of torsion, but their role remains unclear, and a link has not yet been proven.

Due to the high possibility of contralateral torsion and, therefore, the substantial risk of anorchia, there is general consensus among clinicians regarding the necessity of contralateral testicle fixation whenever an orchiectomy after torsion or orchidofixation after detorsion is performed [29–32]. However, some authors do not agree with this policy. Arnbjornson and Mizrahi stated that contralateral orchidofixation is not necessary or advisable [33, 34]. In both trials, no torsion of the contralateral testicle occurred during follow up after surgical scrotal exploration and detorsion of the gonad. The obvious weakness of both studies was a short follow-up period (7 and 6–7 years on average, respectively) and the low number of patients. Surprisingly enough, no long-term, multi-institutional studies have been published to confirm or discredit the necessity of contralateral testicle fixation after unilateral torsion.

When discussing the necessity of healthy testis fixation, one must keep in mind that surgical intervention cannot prevent the testis from torsion in all cases. In 2002, Glabeke *et al.* presented a case report on a patient who underwent orchiectomy due to testicular torsion, and contralateral orchidofixation was carried out at the same time. Ten years later, the patient presented with a torsion of the previously fixed remaining testis, which required orchiectomy and, consequently, led to anorchia [35]. Sells *et al.* published a literature review on secondary torsion after previous orchidofixation [36]. Out of 17 procedures, 15 were performed with absorbable sutures, which the authors suggested might be cause of secondary torsion, and should therefore be avoided for testis fixation. In a 2006 study, Mor *et al.* described eight consecutive cases who suffered secondary testicular torsion, four of whom had ipsilateral and four had contralateral testicular torsion after previous orchidofixation [37]. In those patients, non-absorbable thread was used. The authors highlighted that regardless of the suture material, non-absorbable or absorbable, there is still a risk of secondary torsion.

No recommendations exist on fixation of the contralateral testicle after testicular loss due to trauma or tumor. As a result, the majority of surgeons act according to their experience or preferred therapy. In 1992, Mishriki asked pediatric surgeons and urologists in Great Britain whether they routinely perform orchidopexy in patients with solitary testis who have undergone orchiectomy due to causes other than testicular torsion [29]. Nearly 50% of them admitted that they never perform orchidopexy in such cases, compared to 11% who did it every time. In a study conducted 26 years later, Abdelhalim *et al.* repeated the same survey and found similar results, with only

2% of clinicians stating that they would fix the contralateral testicle after oncological orchiectomy, and less than 10% would perform an orchidofixation in case of testicular trauma [38]. Apart from these questionnaire answers, well-documented evidence on this topic does not exist.

Complications and consequences of surgical intervention on the healthy testis

It is important to consider all of the pros and cons of solitary testis fixation in children, especially the potential side effects of the surgery. Repeatedly piercing the tunica albuginea with a needle may impact the development and maturation of the testicular tissue. In particular, the patient's excretory function and fertility may be impaired. Even so-called "no-touch techniques" such as a dartos pouch, which does not affect the integrity of the tunica albuginea and appears to be less damaging to the gonad, might have negative consequences in the long run. Wound infection and hematoma may also occur after scrotal exploration, which, as the worst case scenario, may result in testis atrophy. However, no data on this topic can be found in the literature.

In 1991, Woitek *et al.* assessed the growth of the solitary testis that was fixed during scrotal exploration for contralateral torsion in childhood [39]. The authors did not find any disturbances in testicular development after fixation. Thus, they suggested that concern about further testicular development should not be a decisive factor in this matter. As most orchidopexies are carried out in childhood for UDT, the effects of this fixation technique have been examined in this population. In 2002, Kozminski retrospectively analyzed 1104 patients who had undergone orchidopexy for UDT that did not affect the tunica albuginea [40]. The results were satisfactory in terms of the testicle size during follow up, and the authors concluded that this confirms proper testicular development. However, it should be noted that the study population consisted of males aged from 1 month to 25 years and included individuals who suffered from torsion as well as those with UDT. Furthermore, it is not yet clear whether a normal testis size is correlated with proper function. Nevertheless, assessing testicular function in patients with UDT is challenging because it is difficult to identify which condition plays a greater role – surgical intervention or cryptorchidism itself.

The hormonal and reproductive function of the fixed testicle in humans remains an uncharted area for clinicians and researchers. Animal models have been used to study these issues. In 2015, Ribeiro *et al.* evaluated the impact of testicular tunical fixation on semen quality and gonadal morphology in three consecutive rat populations: prepubertal, pubertal, and adult [41]. Semen was extracted from the fixed gonads and from the non-fixed counterparts, representing the control group. There were significant differences in the viability and motility of sperm collected from fixed and non-fixed gonads, but a statistically significant difference was only seen in

the adult population. In contrast, morphological changes in the seminiferous ducts, manifesting as decreased thickness and length, were present in both the adult and prepubertal groups. Moreover, these changes were also observed in the non-fixed (contralateral) testicles making up the control group. This suggests that destroying the integrity of the tunica albuginea may be destructive for the contralateral testicle tissue by inducing an autoimmune reaction. In this study, the solitary testicle was not included, but one may assume that the effect on the sperm quality may be exacerbated in cases of monorchism.

Furthermore, monorchism and the timing of testicular loss can, in theory, influence the hormonal and reproductive function of an individual. Grinspon *et al.* assessed 89 patients with congenital or acquired monorchism (divided evenly) by measuring the FSH, LH, AMH and testosterone levels, which were expected to correspond to the reproductive and endocrine function of the single gonad [17]. The results were compared to a group of healthy boys, and no differences in terms of hormonal function (LH and testosterone levels) were observed between the groups. Conversely, the reproductive function, assessed indirectly through the FSH and AMH levels, appeared to be impaired in the monorchism group, becoming even more severe in older patients.

There is little known about the impact of orchidopexy on the reproductive and hormonal function in patients with congenital monorchism, which was summarized by Kehoe *et al.* in 2017 [42].

Summary

Congenital and acquired monorchism have different etiologies, and might also have a different natural history. In particular, the role of the bell clapper anomaly requires further clarification.

Based on current knowledge, it is almost impossible to make definitive recommendations on how to deal with congenital monorchism in terms of contralateral fixation. There is probably no need to perform fixation in all patients, but it is important to identify high-risk patients. The benefits and risks must be weighed in every case.

In the contemporary public health system, young patients with gonadal anomalies are treated by a pediatric surgeon, pediatric urologist or by a urologist. Unfortunately, neither European Association of Urology (EAU) nor European Society of Paediatric Urology (ESPU) have clear guidelines on solitary testicle fixation for various conditions. There is general agreement that the contralateral testis should be fixed in cases of perinatal and postnatal torsion. However, clinical doubt concerning other situations that leave a patient with a solitary testicle, such as trauma, tumor or VTS, remain unresolved.

Last but not least, the impact of the surgical procedure on the maturation and function of the remaining testis should be evaluated in upcoming studies.

Conflict of interest

None declared.

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